Important update on ICD-11 Beta draft proposals

Suzy Chapman | November 7, 2017 | Dx Revision Watch

On November 6, a proposal for significant changes to the ICD-11 concept term, *Postviral fatigue syndrome*, was submitted to the Beta Proposal Mechanism by Dr Tarun Dua.

Dr Dua is a medical officer working on the *Program for Neurological Diseases and Neuroscience, Management of Mental and Brain Disorders, Department of Mental Health and Substance Abuse, World Health Organization.*

Dr Dua is not a member of the *ICD-11 Joint Task Force* but served as lead WHO Secretariat for ICD-11's *Topic Advisory Group for Neurology* which ceased operations in October 2016.

Note: this proposal uploaded by Dr Dua is still at the **"Submitted"** stage - ie, it has not been marked as **"Approved"** or **"Implemented"** or **"Rejected"** nor have any changes to the existing listing for the terms, *Postviral fatigue syndrome*, *Chronic fatigue syndrome*, and *Benign myalgic encephalomyelitis* been inserted directly into the Beta draft, which stands as it did at March 26, 2017 when the terms were restored to the draft.

The proposal submitted by Chapman and Dimmock on March 27, 2017 remains unprocessed by ICD Revision.

Leaving aside the proposal, per se, the content of the rationale that accompanies it, the misconceptions contained within it and the narrow range of studies it relies on, there is a great deal that is odd about this submission. The language is a mash up of report style and peevish colloquial. The author is evidently unfamiliar with the nomenclature used in ICD-11 for the terms currently coded under the concept title, *Postviral fatigue syndrome*: the proposal refers to "Myalgic encephalitis/Chronic Fatigue Syndrome" and "ME/CFS" throughout the submission, whereas the terms classified in ICD-10 and ICD-11 are *"Benign myalgic encephalomyelitis"* and *"Chronic fatigue syndrome"*. The author provides no rationale for combining the terms. The author appears unfamiliar with ICD-11 conventions: ICD-11 does not use acronyms for either concept title terms or inclusion terms; and ICD does not conjoin ICD terms, as in "ME/CFS".

The author has omitted to follow ICD Revision guidance for submitting proposals that involve *"Complex Hierarchical Changes"*: no proposed hierarchical structure for *Chronic fatigue syndrome* and *Benign myalgic encephalomyelitis* has been set out. Is the author proposing *Chronic fatigue syndrome* is elevated to concept title term with *Benign myalgic encephalomyelitis* as the specified inclusion term, or is *Benign myalgic encephalomyelitis* proposed to be included under synonyms or an index term, only? It's not clear.

Whilst the proposal is for deletion of *Postviral fatigue syndrome* from the chapter, *Diseases of the nervous system*, Dr Dua provides no further recommendations for this entity. Does the proposal intend to retire the term? Is *Postviral fatigue syndrome* intended to be retained under synonyms or as an index term under [a relocated] *Chronic fatigue syndrome*? Or do the proposers intend to retain *Postviral fatigue syndrome* but move it to a different chapter location or parent block? No draft *Description* text has been suggested. There is no discussion of whether consideration had been given to creating a new parent class as an alternative to placing under the *"Symptoms, signs or clinical findings of the musculoskeletal system"* block.

Given the imminent finalization of the ICD-11 draft, it is a dog's breakfast of a proposal. Furthermore, it isn't clear whose position this proposal represents. I have asked Dr Dua to clarify whether this proposal represents the position of her WHO department, the *Program for Neurological Diseases and Neuroscience, Management of Mental and Brain Disorder*s; or whether it represents the position of ICD Revision or the Joint Task Force. Has Dr Dua or her department been tasked by ICD Revision to make recommendations or is this proposal unsolicited?

Until clarifications have been provided and until it has been established whether this proposal represents the official, consensus position of ICD Revision or the Joint Task Force, stakeholders and stakeholder organizations are not in a position to submit informed responses.

My recommendation would be to wait until we have obtained those clarifications.

This new proposal, posted yesterday by Dr Dua, proposes to Delete *Postviral fatigue syndrome* from the *Diseases of the nervous system chapter* and relocate "ME/CFS" [sic] to the Symptoms, signs chapter.

Dr Dua's proposal: *"...recommends to remove Myalgic encephalitis [sic]/Chronic Fatigue Syndrome (ME/CFS) [sic] from the nervous system diseases chapter. The rationale for the proposal is lack of evidence regarding any neurological etiopathogenesis of chronic fatigue syndrome. We suggest that ME/CFS [sic] be classified in the Signs and Symptoms Block of the ICD-11 as a child of Symptoms, signs or clinical findings of the musculoskeletal system. The classification in this position according to symptom patterns and severity would be consistent with existing evidence: the syndrome consists of a multitude of symptoms, has an ill-defined pathophysiological etiology, and is a diagnosis of exclusion requiring medical evaluation [1]. When there is sufficient evidence and understanding of the pathophysiological mechanisms, diagnostic biomarkers, and specific treatments, the syndrome can be appropriately classified within the proper block."*

Unless you are registered with the ICD-11 Beta draft for increased interaction with the platform, you won't be able to view Dr Dua's proposal and rationale in the Proposal Mechanism. For ease of access, I am appending a copy of Dr Dua's full proposal and references. I have submitted some comments in which I have requested a number of important clarifications.

ICD-11 Beta Proposal Mechanism

https://icd.who.int/dev11/proposals/f/en#/http://id.who.int/icd/entity/569175314?readOnly=true&action=DeleteEntityProposal&stableProposalGroupId=303c7493-554a-44c8-8e00-bd0c6c4cc6ef

Submitted by Dr Tarun Dua, November 6, 2017

Proposal Status: Submitted

Proposal for Deletion of the Entity Postviral fatigue syndrome

Rationale

Chronic Fatigue Syndrome Proposal

This proposal recommends to remove Myalgic encephalitis [sic]/Chronic Fatigue Syndrome (ME/CFS) from the nervous system diseases chapter. The rationale for the proposal is lack of evidence regarding any neurological etiopathogenesis of chronic fatigue syndrome. We suggest that ME/CFS be classified in the Signs and Symptoms Block of the ICD-11 as a child of Symptoms, signs or clinical findings of the musculoskeletal system. The classification in this position according to symptom patterns and severity would be consistent with existing evidence: the syndrome consists of a multitude of symptoms, has an illdefined pathophysiological etiology, and is a diagnosis of exclusion requiring medical evaluation [1]. When there is sufficient evidence and understanding of the pathophysiological mechanisms, diagnostic biomarkers, and specific treatments, the syndrome can be appropriately classified within the proper block.

ME/CFS is a Syndrome of a Constellation of Symptoms and Signs

The predominant symptom of those with ME/CFS present is severe fatigue, a manifestation of skeletal muscle dysfunction. In addition, these patients may report pain, cognitive symptoms, myalgia, impaired memory or concentration, gastrointestinal problems, headaches, and arthralgia. Less commonly, individuals report dizziness, nausea, anorexia, and night sweats. Signs include tender lymph nodes and a sore throat [2].

Epidemiological and Pathophysiological evidence is limited, conflicting, and does not support ME/CFS as a disease of the nervous system or with a principally neurobiological underpinning

The underlying pathophysiological basis of ME/CFS remains unclear. This is in part due to methodological limitations in epidemiological studies given variability in case definitions [1, 3-5]. Prevalence and incidence of ME/CFS varies greatly across age, gender, ethnicity, socioeconomic strata and country, without clear explanation of the differences [6-12].

Much of the study to date on biological mechanisms has been focused on the central nervous system and immune systems with conflicting results [2]. Very limited evidence points to the nervous system as the site of pathology [13], with no clear patterns of CNS involvement [14-17].

Though serotonergic and cortisol responses have been abnormal, no consistent alterations in the function of the hypothalamo-pituitary-adrenal axis, stress hormone pathways, or immune system have been identified among those with ME/CFS [18-20]. Further, research examining metabolic, sleep or psychological models of the disease is also inconclusive. The etiology and pathogenesis of CFS are hypothesized to be multi-systemic, multifactorial and require predisposing (genetic, lifestyle), precipitating (infection, psychological stress) and perpetuating factors (psychosocial processes)[2]. For example, it has been demonstrated that a stereotyped syndrome of disabling fatigue, musculoskeletal pain, neurocognitive symptoms, and mood disturbance occurred after viral infection by infection with Epstein-Barr virus (glandular fever), Coxiella burnetii (Q fever) and Ross River virus (epidemic polyarthritis) [21].

Given the persistent lack of an understanding of etiology of ME/CFS, a European database has been established to examine biomarker research for clinical use; the ME/CFS EUROMENE database has confirmed the presence of heterogeneous evidence regarding neurological, immune and metabolic markers that vary by gender, and hypothesize a multifactorial syndrome with environmental and immunological factors as the biological basis of ME/CFS [22].

The definitions and classification of ME/CFS emphasize the need for a systems-based approach.

There are at least 20 case definitions of ME/CFS, and no systematic evidence was present that any definition “specifically identified patients with a neuroimmunological condition” [1]. Definitions including CDC 1994-Fukuda [23], ME-International Consensus Criteria (ME-ICC)[24], and the 2015 Institute of Medicine report on ME/CFS proposing the new name of systemic exertion intolerance disease (SEID) [3- 5], further highlight that there is a changing understanding of the nature and cause of the illness.

These changing definitions and classifications further reflect the need for a systemic characterization of the illness while recognizing the impact on multiple organ systems, and that varying types of exertion (emotional, cognitive, or physical), are characteristics of the syndrome. There are thirteen synonyms in the Foundation layer; only reinforce the opinion that this is a very imprecise disorder. It is interesting to note that Akureyri (mentioned twice) is a city in Iceland and again to mention Iceland disease and Icelandic disease! All this has to be cleaned and taken out in the new position of CFS. The body site is NOT entire brain and a virus does NOT cause it.

The treatment of ME/CFS centers around graduated exercise and psychologically based treatments.

It is also important to recognize that diagnosis and appropriate referral for treatment are established and accessible in a variety of settings (such as primary care, emergency departments, mental and behavioral health clinics, medical subspecialties (infectious diseases, cardiology, and rheumatology as well as pediatrics) [25, 26]. The most robust evidence of symptomatic treatment with observed functional benefits is limited to Cognitive behavioral therapy and graded exercise [27-31].

ME/CFS is thus not a disease of the nervous system. It should be categorized in the Signs and Symptoms chapter given the lack of clear evidence pointing to the etiology and pathophysiology of this syndrome until evidence to organ placement is clarified in years to come.

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